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Case Report

Choreoathetotic syndrome following cardiac surgery☆,☆☆,★,★★

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Abstract Movement disorders following heart surgery are very unusual. Post-pump chorea is mainly a pediatric complication of heart surgery, typically manifesting after a latent period of normality and is usually related with long extracorporeal circulation time and deep hypothermia. We report a 73-year-old woman, without risk factors predisposing to paroxysmal movement disorders, presenting acute choreoathetoid movements 5 days after aortic valvular replacement with normal extracorporeal circulation time and perioperative normothermia.

1. Introduction

We present a case of post-pump choreoathetotic syndrome after aortic valvular replacement in a 73-year-old female. We aim to highlight this uncommon neurological disorder which is mainly a pediatric complication, typically manifesting after a latent period of normality. Few cases of adult post-pump chorea have been described, and they are all related to long extracorporeal circulation (ECC) time and deep hypothermia [1].

2. Case report

A 73-year-old female suffering from mild hypertension and hypothyroidism underwent heart surgery for valvular aortic replacement because of severe aortic valve stenosis. Anesthesia was induced with midazolam, ketamine, sufentanil, cisatracurium and a low dose of propofol. The maintenance of anesthesia was made with sevoflurane. One pack of red blood cells was given during cardiopulmonary bypass for hemoglobin at 8.1 g/dL. Norepinephrine (maximum dosage was 0.05 μg/kg per minute) was used after cardiopulmonary bypass to

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maintain adequate blood pressure for a short period of time. ECG, hemodynamics and cerebral oxymetry remained normal throughout the all procedure. The aortic clamping time was 34 minutes and no perioperative hypothermia was signaled. The total ECC time was 68 minutes. There was no hyperglycemia during surgery. No noticeable incident was observed in the early post-operative period and during hospitalization.

She presented to the emergency department ten days after surgery because of occurrence, since 5 days, of acute choreoathetoid movements involving mainly the lower limbs and the face with orofacial dyskinesia and blepharospasm. Neurological examination also revealed bradyphrenia, dysphagia and psychomotor impairment. A careful history ruled out rheumatic fever, streptococcal infection and family history of degenerative disease or autosomic dominant or recessive inheritance. A previous exposition to neuroleptic agents was considered as these drugs can cause tardive dyskinesia (chorea) but she had never taken such drugs. Because of the acute onset of symptoms, it was imperative to consider metabolic, endocrine, post-infectious or auto-immune condition: blood sugar and thyroid function were normal, anti-nuclear and anticardiolipin antibodies were negative and excluded systemic lupus erythematosus or anti-phosphilipid antibody syndrome. Anti-streptolysin O titer was tested to measure antibodies against streptolysin O and ruled out streptococcal infection. Despite age at onset made improbable the possibility of Wilson disease or acanthocytosis, ceruloplasmin, copper, creatine kinase were tested and found to be normal.

Brain magnetic resonance examination showed leukoaraiosis but no pathological findings in the basal ganglia. EEG showed a slow activity without focal abnormalities. Cerebral positron tomography using [18F] fluorodeoxyglucose (FDGC-PET) showed generalized hypometabolism within the occipital, temporal and inferior parietal cortex. Analysis of cerebrospinal fluid (CSF) was negative.

Based on non-relevant medical history including genetic disorder, normal laboratory findings for alternative diagnoses and normal brain MRI, the diagnosis of post-pump chorea (PPC) was retained. Even though a slowly progressive clinical improvement was observed after treatment with tiapride 50 mg 3 times a day, residual choreo-athetoid movements persist at seven months of follow-up.

Clinical features are illustrated in Videos 1 and 2. To access video component, click on the images in supplementary files.

3. Discussion

Choreoathetoid movements following open-heart surgery, the so called PPC, is a rare condition affecting almost exclusively children or adolescents undergoing cardiac surgery and it has rarely been reported in adults [2-4]. PPC has also been described in some adults after pulmonary endarterectomy with deep hypothermia and cardiopulmonary bypass [5,6].

The acute onset of PPC occurs 2–14 days after surgery, typically after an asymptomatic period [7]. The clinical presentation appears similar in children and adults. Potential risk factors have been reported, such as a longer aortic clamp time during surgery, the time on ECC [8], a preoperative cyanosis, a deep hypothermia longer than 60 min, an alpha-stat pH management, a pre-existing developmental delay [3]. Medlock et al. postulated a strong association between circulatory arrest, deep hypothermia and PPC [8], but occurrence of this syndrome without these conditions has been described in children [8]. However, to our knowledge, all the reported cases in adults occurred with deep hypothermia or longer aortic clamp or ECC time, contrary to our patient in whom cardiopulmonary bypass was the only risk factor, showing that hypothermia is not a necessary condition for PPC. The total ECC time observed in our patient is not excessive and is significantly lower than the median value observed in a series of 5 patients who developed chorea in the postoperative period after pulmonary endarterectomy [6] (137 minutes ±26) and in a 77-year-old male who underwent aortic valve replacement and coronary artery bypass grafting (144 minutes) [3]. This highlights that a long time on ECC is not essential for occurrence of PPC.

Choreoathetosis can involve the four limbs with orofacial dyskinesia, dysphagia, loss of tone and impairment of speech. Oculomotor apraxia is another clinical feature but less observed [9]. The predominant involvement of the lower limbs and the face is a main feature of this disorder. Detailed medical history, accurate drug history, laboratory investigations and neuroimaging are useful to exclude other causes of chorea.

The pathogenesis of PPC remains unclear, but the literature suggests it may be the result of a biochemical or microembolic phenomena affecting basal ganglia. Indeed, hypothermia could lead to lactic acidosis, resulting in a damage of striatum [2]. The hypothesis of an increased blood viscosity induced by ECC has also been suggested [2]. Surie et al proposed the hypothesis that faster rewarming induces an increase in the cerebral metabolic rate for oxygen surpassing the recovery of the cerebral blood flow [6]. However, the occurrence of PPC with normothermia in our case suggests a more complex physiopathological pathway. The higher frequency observed in children remains yet to be elucidated.

FDGC-PET study argues in favor of a vulnerability of basal ganglia since bilateral basal ganglia deep hypometabolism was shown in a 52-year-old woman presenting a choreoathetoid syndrome following heart surgery for thoracic aortal aneurysm dissection [2]. Data of FDGC-PET study in our case suggest metabolic change facilitating the occurrence of PPC. We therefore suggest that multifactorial mechanisms may contribute to the development of choreoathetotic syndrome as a complication of cardiac surgery.

The course of PPC is variable; it can be transient or severe and irreversible, possibly associated with cognitive disturbances and severe disability [7,8]. Improvement is variable and can be observed early (two days) [9] or after 2 months [8]. The available therapy is only symptomatic (neuroleptics drugs). This is why we used tiapride (50 mg three times a day with slowly decrease), a blocker of dopamine receptors, with a favorable outcome. Excellent outcome in a 77-year-